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Case report - Esophagus

Esophageal cancer complicated with azygos continuation
of the inferior vena cava

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Abstract

Neoadjuvant (preoperative) therapy and combined modality therapy have become focuses of interest in the effort to prolong survival and to reduce recurrence rates in patients with esophageal cancer. Staging of the tumor is a critical step in establishing which therapeutic option is appropriate. Once surgical management is advocated, adequate medical imaging is crucial in determining individual anatomical variations. In this communication we report a case of a patient with azygos continuation who underwent chemoradiotherapy with successful downstaging of tumor status from T_{3–4} to T₀ and a nodal status from N₁ to N₀ as evaluated by medical imaging and who then proceeded to curative surgical resection. This case highlights the potential ability of radiological techniques to confirm both anatomical variations and responses to neoadjuvant therapy.

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Therapy for esophageal carcinoma is influenced by the knowledge that in most of these patients, local tumor invasion or distant metastatic disease precludes cure. Appropriate selection of patients with esophageal carcinoma for surgery or chemo-radiotherapy depends critically on accurate staging. Multimodality therapy (chemotherapy and radiation therapy) followed by total esophagectomy has been reported to improve survival compared with esophagectomy alone, with an approximate doubling of survival [1]. In addition, in patients with locally advanced disease the potential benefit of the preoperative chemoradiation becomes more evident; many patients have the stage of their disease lowered and become eligible for complete resection.

In selected cases, some anatomical variations and congenital malformations may adversely influence surgical manipulation. Certain anatomical structures – that are normally resected in order for establishing an R₀-resection – must be spared if an anomaly is suspected. With respect to esophageal malignancies, this is the very case with

the azygos system. The azygos vein is normally resected during esophagectomy. Ligation of the azygos vein, however, in the presence of an azygos-caval anomaly abolishes venous drainage from the lower half of the body, leading to venous hypertension and ultimately to death. Therefore, as we present that in this communication, medical imaging is not only essential in staging of the tumor, but also crucial in determining such anomalies prior to surgical intervention.

2. Case report

A 52-year-old white male with a history of epilepsy presented with dysphagia for semi-solids and weight loss of 10 kg during a 3-month period. He smoked three packs of cigarettes daily and admitted to intermittently heavy alcohol ingestion. His current medication was tiapride; he had no family history of note. He had no other symptoms and physical examination was unremarkable.

The plain chest radiograph was also completely normal. Computed tomography (CT) of the chest and abdomen revealed an advanced mid-esophageal mass with extensive periaortic and aortic window lymphadenopathy. Scans proved invasion of the trachea. The images also showed

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a dilated azygos vein and arch in their paravertebral course associated with non-visualization of the intrathoracic inferior vena cava (IVC) (Fig. 1). Sections through the upper abdomen showed absence of supra-renal IVC with dilated azygos vein. The cardiac chambers, lungs, liver, and hepatic veins were normal, and there was no situs inversus. Taken together, an azygos continuation of the IVC was suspected. However, CT scans also revealed invasion of the dilated anomalous azygos vein.

An upper gastrointestinal (GI) gastrograffin contrast radiograph series showed a mid-esophageal filling defect. The patient had undergone esophago-gastro-duodenoscopy (EGD), which revealed a mass at 27 cm from the incisors. A biopsy from the lesion disclosed squamous cell carcinoma. Bronchoscopy demonstrated signs of external compression of the trachea with no mucosal lesion. By utilizing complex medical imaging, the locally advanced tumor was staged as T₄N₁M₀ and was deemed unresectable by both oncological and technical means.

The patient then underwent neoadjuvant chemoradiotherapy, which he tolerated well without complication. Chemotherapy consisted of two cycles of carboplatin and 5-fluorouracil, administered 3 weeks apart. Three-hundred mg of carboplatin was infused on day 1 as a single dose. A total of 1000 mg of 5-fluorouracil was given as a continuous infusion on days 1–5. External beam radiation therapy was concurrently administered during chemotherapy. The patient received a total dose of 27.5 Gy in 2.5 Gy daily fractions.

After neoadjuvant chemoradiation, repeat CT scans of the chest and abdomen demonstrated total reduction in the size



Fig. 1. The CT scan demonstrates a mid-esophageal mass with the invasion of the trachea (arrow) and the presence of mediastinal lymphadenomegaly. Images also prove dilated azygos vein and arch in their paravertebral course (asterisk). In conjunction with an undetectable inferior vena cava, the diagnosis of azygos continuation was made.



Fig. 2. Chest CT image proves a down-staged mid-esophageal mass after neoadjuvant chemoradiotherapy (arrow). The tumor and lymphadenomegalies are unappreciable. Dilated azygos vein and arch (asterisk) along with an absent intrathoracic inferior vena cava reconfirm azygos-caval anomaly.

of the tumor and showed no evidence of metastatic disease (Fig. 2). Both trachea and azygos vein were free of neoplastic invasion. The smaller peritumoral and periaortic nodes were no longer visible. CT scans have also reconfirmed the presence of azygos continuation – a congenital malformation that a general surgical team scarcely encounters with. At repeat EGD no evidence of his primary tumor could be found. Bronchoscopy revealed neither endotracheal, nor endobronchial propagation of the carcinoma. Taken together, the patient was successfully down-staged.

He then proceeded to successful Ivor-Lewis esophagectomy, sparing the azygos vein, with an uneventful post-operative recovery. Pathology of the resected specimen found residual primary tumor in the esophagus (histologically: non-keratinizing squamous cell carcinoma) and no evidence of malignancy in any of the six lymph nodes examined.

At follow-up 12 months later the patient was asymptomatic with no evidence of recurrent disease clinically or on CT.

3. Discussion

Esophageal cancer is notorious for its aggressive biologic behavior; it infiltrates locally, involves adjacent lymph nodes, and metastasizes widely by hematogenous spread. Unfortunately, most patients still present with locally advanced (stage T₃ and/or N₁) disease. Prognosis and outcomes are determined by stage. Our responding patient

with a locally advanced disease underwent preoperative neoadjuvant chemoradiation. He was successfully down-staged, and then proceeded to curative surgical resection. He returned to good health and has returned to work. He remains free of disease 12 months after the initiation of therapy.

Preliminary evidence suggests that multimodality treatment with radiation therapy plus chemotherapy followed by resection is superior to any therapy alone. Chemoradiotherapy is used preoperatively to treat micrometastases and to reduce the size of the tumor to improve resectability rate. Such a regimen should increase the R_0 resection rate in patients with locally advanced tumors and in patients with tumors at unfavorable locations, should reduce the rate of local and distant recurrences, and should thereby increase the chances for long-term survival.

In performing esophagectomy, the azygos vein and the thoracic duct are resected along with the primary specimen. Developmental anomalies of the IVC are relatively rare, occurring in <1% of patients with congenital heart disease [2]. When developmental interruption of the infrahepatic IVC occurs, venous return from the lower body is via the azygos or hemiazygos venous system. A high incidence of associated congenital cardiac malformations, errors in abdominal situs, asplenia, and polysplenia were subsequently reported. This anomaly was first described by Stark in the mid-19th century [3]. Ever since a couple of cases have been published, however this is the first report in which successful esophagectomy was performed in this rare disorder. The anomaly was detected in the past by either inferior vena cavography, transfemoral cardiac catheterization, or exploratory thoracotomy. With the advent of CT scan and magnetic resonance imaging (MRI), the normal anatomy of the azygos vein and its abnormalities are well visualized without the need for invasive procedures.

In the case of our patient with esophageal cancer invading adjacent structures, the anomalous azygos vein was not free of neoplastic involvement at presentation. Because of the unfortunate association of his malignancy

with an azygos-caval anomaly, this simple finding had furthered the locally advanced oncologically unresectable mid-esophageal tumor to be deemed technically unmanageable by surgical means. Having undergone preoperative chemoradiation, however, the patient became free of invasion of the azygos system – the only vessel through which veins from his lower half of the body drained. Therefore in such cases, just as presented in this brief clinical report, neoadjuvant therapy is ultimately essential not only in down-staging a tumor, but making a neoplastic lesion both oncologically and technically resectable.

Moreover, failure to detect the presence of azygos-caval anomaly may be lethal for patients undergoing esophageal or other thoracic surgery, as its ligation abolishes venous drainage from the lower half of the body, resulting in venous hypertension and death. Invasive procedures can be avoided if the diagnosis is suspected and a CT scan or MRI performed.

Versatile radiological techniques have emerged that are essential in staging and re-staging the disease after preoperative treatment, and verifying responses to neoadjuvant therapy. Many-sided medical imaging is also imperative in confirming anatomical variations. Taken together, hand-in-hand collaboration of the surgical team with oncological and radiological units is crucial in giving the patient the best available treatment utilizing evidence-based medicine.

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